previous studies with ill subjects, children with diabetes were more likely to include the pancreas. They were also significantly less likely to draw the brain or stomach than the healthy group. The drawings of the diabetic children who did not include the brain were not inferior in any other respect to those of their healthy controls. Their responses to questions about the function of the body organs also indicated that they knew they had a brain and why. Unfortunately, it was not possible to go back and ask children why they had originally omitted to draw the brain, but in the one case where this was possible, the girl (aged 15 years) said that what was in her head seemed of secondary importance to what was in her 'insides'. Perhaps this might lead one to speculate that diabetic and healthy children assign differences in value to various body parts. These data suggest that although there are few absolute differences between diabetic and healthy children in knowledge of their bodies, there may well be differences in attitude so that diabetics see some parts of their bodies as more or less important than healthy children.

Diabetic and control children attributed diabetes to different things. The control group favoured explanations like heredity, and the result of eating too much sugar. In contrast, paediatricians might feel heartened to learn that the types of explanations offered by the diabetic child were consistent with those generally offered by the medical profession. Only a small number apportioned any self-blame for the illness—challenging the established view that chronically sick children tend to blame themselves.⁶

The children included in this study all seemed to

be managing their disease and its impact on their lives relatively well. These results may not, however, obtain for other groups of sick children, such as those with diseases perceived to be more threatening, or those with less supportive home backgrounds. Nevertheless, the data do suggest that it is possible to suffer a relatively threatening chronic illness during childhood, and for this to have few repercussions for the development of the child's knowledge of the body.

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Megacystis-microcolon-intestinal hypoperistalsis syndrome

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SUMMARY Two neonates with megacystis-microcolon-intestinal hypoperistalsis syndrome are described. Both were boys. The main manifestation was functional obstruction of the urinary and gastrointestinal tracts. While there is no explanation as yet for the aetiology and pathogenesis, the diagnosis is simple, the treatment symptomatic, and the outcome fatal.

Megacystis - microcolon - intestinal hypoperistalsis syndrome was first reported by Berdon.¹ Neonates with this syndrome have a distended urinary bladder and hypoperistalsis throughout the entire gastrointestinal tract. Most of the reported cases have been girls.¹⁻⁴ We report two boys with this rare syndrome and discuss its diagnostic and treatment implications.

Case reports

Case 1. This boy was born at term weighing 3800 g. He failed to pass meconium for 48 hours and developed abdominal distention and bilious vomiting. At operation at the age of 2 days a very large flaccid bladder and a shortened small bowel containing thick inspissated meconium were found. The colon was narrow and empty. There was also malfixation of the midgut. Seromuscular biopsies of the colon and small bowel showed normal ganglion cells in the myenteric plexus. A Roux-en-Y Bishop-Koop type enterostomy was performed above the inspissated meconium. After the operation the bladder had to be emptied by repeated catheterisation. The ileostomy did not work and abdominal distention recurred. Intravenous urography and retrograde cystography showed severe bilateral hydronephrosis and hydroureters and a large flaccid urinary bladder without vesicoureteric reflux. The patient was operated on again at the age of 6 days. A segment of infarcted small and large bowel was resected and an end jejunostomy, transverse colostomy, and suprapubic cystostomy were performed. All attempts at oral feeding failed because of repeated bouts of abdominal distention, and the infant was maintained on long term total parenteral nutrition. The patient was referred to our department at the age of 5 months, still weighing 3500 g. A barium swallow at that time showed a distended stomach and a 20 cm long small bowel, ending at the jejunostomy site. Complete absence of peristalsis was noted and the contrast material was present in the oesophagus, stomach, and jejunum 8 hours later (Fig. 1) as well as after three days. The patient died shortly thereafter owing to fulminant urinary sepsis.

Case 2. A 34 year old woman underwent sonographic evaluation for polyhydramnios in month 8 of pregnancy. This showed a fetus with a large intra-abdominal cystic mass (Fig. 2). A boy was born at term, weighing 2800 g. He had a notably distended abdomen due to a large bladder that contained over 500 cc of urine. Decompression of the bladder relieved the abdominal distention and produced a flaccid abdominal wall suggesting prune belly syndrome. A barium enema showed a microcolon and most of the barium failed to evacuate over the next 24 hours. Rectal mucosal biopsies showed abundant ganglion cells in the submucous plexus. Radiographs showed normal kidneys and a large flaccid urinary bladder without bladder neck obstruction or vesicoureteric reflux. Attempts to feed him caused abdominal distention. A transperitoneal suprapubic cystostomy was performed at the age of 2 weeks. Postoperatively, the patient had to be maintained on

longterm parenteral nutrition. There was progressive deterioration of renal function with increasing dilatation of the upper urinary tracts. The infant died of renal failure at the age of 5 months.

Fig. 1 Case 1—barium swallow showing a dilated stomach with poor emptying after 8 hours.

Fig. 2 Case 2—fetal ultrasound scan showing a large cystic mass (CM) in the patient's abdomen (AF=amniotic fluid).

Discussion

There are only 20 patients with megacystis-microcolon-intestinal hypoperistalsis syndrome in the published reports, and almost all were born at term and were girls.¹⁻⁵ The syndrome encompasses anomalies of two organ systems. In the gastrointestinal tract, shortening and fixation are noted, with a microcolon and diffuse hypoperistalsis. Considerable postnatal abdominal distention with bilious vomiting resembling the clinical picture of mechanical intestinal obstruction called for an early exploratory laparotomy in most reported cases. In the urinary system, a large bladder without bladder neck obstruction is described. In some cases this is accompanied by hydronephrosis and hydroureters.

The pathogenesis of the syndrome remains obscure with no satisfactory explanation for the absence of peristalsis and the flaccid bladder. This syndrome may be a variant of a group of diseases that affect the motility of the gastrointestinal tract. These come under the heading of idiopathic intestinal pseudo-obstruction.⁶ In these disorders ultrastructure examination of the ileum and the urinary bladder showed vascular degenerative changes in the smooth muscle cells.^{5,6} Both patients described here were boys. This is important when the differential diagnosis is considered. Prune belly syndrome predominately affects boys, but neonates with prune belly syndrome have no hypoperistalsis of the gut. In our second patient, decompression of the bladder caused abdominal wall flaccidity that later disappeared.

The megacystis-microcolon-intestinal hypoperistalsis syndrome may be accurately diagnosed by abdominal sonography, cystography, and a barium

enema. A combination of an enlarged bladder without bladder neck obstruction together with a typical 'microcolon' with no radiological evidence of mechanical intestinal obstruction is diagnostic, and should prevent an unnecessary exploration. This was the case in the second patient described. This syndrome was lethal in all the reported patients and only two patients survived for more than one year.^{2,4} Treatment was symptomatic with long term total parenteral nutrition and adequate drainage of urine. Attempts to induce intestinal peristalsis by pharmacological means have so far failed to improve the poor outcome of the disease.³

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Haemophilia and T lymphocyte subsets

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SUMMARY Number of circulating lymphocytes and T cell subsets were assessed in 13 boys with severe haemophilia and 12 age matched controls. There was no significant difference between the two groups. This conflicts with previous findings in adults where lymphopenia and changes in T cell subpopulations have been found frequently.

Lymphopenia with reversal of the ratio of T cell subpopulations defined by the monoclonal antibodies OKT4 and OKT8 (or their equivalents) are constant findings in the acquired immune deficiency syndrome (AIDS),¹ and have been taken as an indicator of latent AIDS when found in asymptomatic subjects.² The disease is thought to be transmissible in blood products, and screening of groups at risk has led to the discovery of the characteristic